DOCUMENT RESUME

ED 406 787 EC 305 488

AUTHOR Bellinger, Diane; And Others
TITLE Fragile X Syndrome in Males: Diagnostic, Behavioral, and Educational Implications.
INSTITUTION Lynchburg Coll., Va.
PUB DATE [95]
NOTE 27p.
PUB TYPE Information Analyses (070)
EDRS PRICE MF01/PC02 Plus Postage.
DESCRIPTORS Autism; Behavior Patterns; Clinical Diagnosis; DNA; Drug Therapy; Educational Needs; Failure to Thrive; *Genetics; Heredity; *Intervention; Language Acquisition; Language Impairments; Learning Disabilities; Males; Medical Services; *Mental Retardation; *Multiple Disabilities; Sensory Integration; Special Health Problems; *Symptoms (Individual Disorders)
IDENTIFIERS *Fragile X Syndrome

ABSTRACT

This paper reviews the research on fragile X syndrome, the second most common cause of mental retardation related to chromosomal anomaly. It notes that far more males than females are affected by the fragile X syndrome, which typically results in craniofacial changes, delays in growth and development, speech/language difficulties, and cognitive deficits including mental retardation or learning disabilities. Topics reviewed include: diagnostic methods (direct DNA analysis is now preferred to cytogenetic analysis or DNA linkage studies); inheritance patterns (which explain the predominance of males); physical and cognitive characteristics; medical conditions associated with the condition (seizure disorders, motor coordination deficits); behavioral characteristics (hyperactivity, gaze avoidance); and speech/language characteristics (perseveration, cluttering, and echolalia). Other topics discussed are a possible relationship between autism and fragile X syndrome, whether language problems characteristic of the syndrome are due to qualitative or quantitative differences in language development, and the common presence of hypersensitivity. Interventions reviewed are organized into medical (especially drug therapy), and educational, which include classroom modifications, training in self-management skills, curricular modifications, and sensory integration therapy. (Contains 35 references.) (DB)

****
* Reproductions supplied by EDRS are the best that can be made *
* from the original document. *
****

U.S. DEPARTMENT OF EDUCATION
Office of Educational Research and Improvement
EDUCATIONAL RESOURCES INFORMATION
CENTER (ERIC)
☐ This document has been reproduced as received from the person or organization originating it.
☐ Minor changes have been made to improve reproduction quality.
• Points of view or opinions stated in this document do not necessarily represent official OERI position or policy.

ED 406 787

Fragile X Syndrome in Males: Diagnostic, Behavioral, and Educational Implications

Diane Bellinger
Appomattox County (VA) Public Schools

Helen Rucker
Lynchburg City (VA) Public Schools

Edward A. Polloway
Professor of Special Education
Lynchburg College

PERMISSION TO REPRODUCE AND DISSEMINATE THIS MATERIAL HAS BEEN GRANTED BY
E. Polloway
TO THE EDUCATIONAL RESOURCES INFORMATION CENTER (ERIC)

EC 305488

Running Head: Fragile X

Address Correspondence to: Edward A. Polloway, Ed.D.
School of Education and Human Development
Lynchburg College
1501 Lakeside Drive
Lynchburg, VA 24501
polloway@hopwood.lynchburg.edu

BEST COPY AVAILABLE

Abstract

This paper provides an overview of fragile X syndrome, the second most common cause of mental retardation related to chromosomal anomaly. The history, diagnostic methods, and characteristics of fragile X syndrome are discussed, as well as educational and medical interventions. Although both males and females can be affected by fragile X syndrome, significantly more research has centered around affected males. Thus this paper primarily addresses that population. Perspectives are provided regarding various intervention strategies.

Fragile X Syndrome in Males:

Diagnostic, Behavioral, and Educational Implications

Fragile X syndrome is second only to Down syndrome as an identified cause of mental retardation related to chromosomal abnormality. The term "fragile X" describes an area on the long arm of the X chromosome which appears to be suspended by a thread from the chromosome itself (Rogers & Simensen, 1987). This chromosomal abnormality typically results in craniofacial changes, delays in growth and development, speech/language difficulties, and cognitive deficits including mental retardation or learning disabilities. In many persons, autistic-like behaviors are also present.

In 1943, Martin and Bell described a British family in which there were 11 males in two generations who had the clinical features of fragile X syndrome and consequently, this syndrome has been commonly referred to Martin-Bell syndrome (de la Cruz, 1985). In 1969, cytogenetic research conducted by Lubs sparked a renewed interest in fragile X syndrome. He observed that distal constrictions on the long arm of the X chromosome were present in four affected males over three generations. However, Lubs' (1969) work was not duplicated until 1977, when researchers in Melbourne reported eight families who demonstrated similar clinical and cytogenetic findings (Harvey, Judge, & Wiener, 1977).

Prevalence figures suggest that approximately 1 in 1000 to 2000 males have fragile X syndrome (Bodurtha, Tams, & Jackson-Cook, 1992). However, utilization of direct DNA analysis, which yields more reliable, conclusive results, may result in increased identification of affected males, as well as carrier females, in the future.

Due to the prevalence of males affected with fragile X syndrome, this paper will focus mainly on males. The paper also describes the large variance in cognitive and behavioral characteristics of individuals with fragile X as well as with recent research on possible links between autism and fragile X. Special attention will be given to emerging perspectives on interventions appropriate for children and adolescents with fragile X syndrome.

Diagnosis and Inheritance Patterns

Diagnostic Methods

The physical features of fragile X syndrome may manifest in varying degrees, and often are not discernible until after childhood. Historically, clinicians proceeded with cytogenetic analysis (for formal diagnosis) only after several family members showed evidence of the clinical findings associated with the syndrome. Cytogenetic analysis requires that a small amount of blood be drawn and subsequently analyzed, with the intent of detecting fragile sites on the X chromosomes. One major limitation of cytogenetic testing is that at least 20 percent of males who are known to carry the fragile X gene do not show evidence of these fragile sites. Additionally, only 55% of obligate carrier females show evidence of fragile sites, with those present in only a small number of cells (Rousseau et al., 1991).

DNA linkage studies may be quite useful among families in which one or more members are affected with fragile X syndrome. In these studies, DNA segments of family members are compared with those of the affected individual(s). However, while this

analysis yields helpful information in some cases, it does not always confirm or rule out the presence of a fragile site (Schopmeyer & Lowe, 1992).

A major victory in the quest for an accurate diagnosis of fragile X syndrome came when the fragile X gene was located by several research groups (e.g., Rousseau et al., 1991; Sutherland et al. 1991; Verkerk et al., 1991). Consequently direct DNA analysis is now used for diagnostic purposes and is both more reliable and more conclusive than either cytogenetic analysis or DNA linkage studies. Additionally, it can be successfully utilized in both prenatal and postnatal analyses. Through this type of testing, female carriers and transmitting males can be identified and the fragile X mutation can be detected prior to birth (Finucane, McConkie-Rosell, & Cronister, 1993).

Inheritance Patterns

A woman is considered a carrier of the syndrome if the fragile X gene is present on one of her two X chromosomes. Because a woman has two X chromosomes, typically she may not exhibit the features of fragile X syndrome since the effects are "concealed" by the healthy gene on the other X chromosome. Each offspring of a carrier mother has a 50% chance of inheriting the fragile X gene. Because males have only one X chromosome in addition to one Y chromosome (the latter of which always comes from the father), consequently, when a male inherits the the fragile X gene, he will usually be affected. Males who have the fragile X gene pass it to all of their daughters; however, the gene is not passed to their sons, who inherit the Y chromosome instead (Bodurtha et al., 1992). Genetic counseling provides families with information about inheritance, diagnosis, and characteristics associated with fragile X syndrome.

Characteristics

A range of characteristics may be associated with fragile X syndrome in males. Commonly described patterns associated with the syndrome have been found in the physical, cognitive, behavioral, and communication domains.

Physical and Cognitive Characteristics

Children born with chromosomal abnormalities in general often exhibit a number of general characteristics, including low birth weight, failure-to-thrive, and various physical anomalies which may affect the development of body systems. Fragile X children generally experience increases in weight, height, and head circumference as they get older. However, as weight and height become proportionate with one another, head circumference often increases significantly, resulting in macrocephaly (Bregman, Dykens, Watson, Ort, & Leckman, 1987). Other general physical characteristics of affected males include: macroorchidism (enlarged testes) after puberty; low-set, protruding ears; high, prominent forehead; and protruding jaw.

Mental retardation is frequently present in varying degrees ranging from severe retardation to borderline levels of intelligence. While females affected with fragile X syndrome may have some of the same craniofacial features as males, their cognitive differences, when present, are more consistent with a diagnosis of learning disabilities than with mental retardation. Math difficulties, language delays, sensory integration problems, and attention deficits are common occurrences (Schopmeyer & Lowe, 1992).

Medical Conditions

According to Bregman et al (1987) and Hagerman (1995), approximately 15% of children with fragile X syndrome have a seizure disorder, which often begins in early childhood. These seizures range from grand mal to brief absence episodes. Chronic otitis media (middle ear) infections and sinus infections are also common, the former of which may result in hearing loss and subsequent delays in language development. Approximately 3% of fragile X children have cleft palate, which can also impact speech and language development. Visual problems are manifested in approximately 50% of fragile X patients, with strabismus presenting in 30% of cases. Other common conditions among those affected with fragile X syndrome include: connective tissue dysplasia, which leads to joint hyperextensibility; hypotonia (floppiness); overall fine and gross motor coordination deficits; as well as difficulty with sensory integration.

Behavioral Characteristics

According to Levitas (1995), many babies with fragile X are considered affectionate, while others tend to move away from hugs, avoid eye contact, shy away from being held, and may not return smiles. These children have a preference for atypical toys, and many taste or smell objects rather than looking at or touching them.

Many affected individuals exhibit specific behavorial problems including: hyperactivity, attention deficits, frequent repetition of the same posture, movement, or form of speech; gaze avoidance; and, at times, aggressive and self-injurious behaviors (Brown et al., 1982; Hagerman, Smith, & Mariner, 1983; Mattei, Mattei, Aumeras, Auger, & Giraud,

1981; Turner, Brookwell, Daniel, Selikowitz, & Zilibowitz, 1980). Early childhood features have been noted to include poor eye contact in 90% of the cases and tactile hypersensitivity. Additionally, these children have difficulty making transitions and often have difficulty with processing and responding to environmental stimuli (Schopmeyer & Lowe, 1992).

Speech and Language Characteristics

Scharfenaker (1995) reported that speech and language characteristics vary among individuals affected with fragile X syndrome, but generally they may include: the repetition of a response over and over (perseveration); rapid vacillation of speech rate with repetition of sounds, syllables, and words (cluttering); poor topic maintenance; inappropriate syntax; and a high receptive vocabulary. Echolalia (i.e., echoing others' voices) and palilalia (i.e., repetition of statements with increasing speed and reduced loudness) have also been noted to occur with frequency among this population (Newell, Sanborn, & Hagerman, 1983).

According to Wolf-Schein et al. (1987), the specific language problems associated with fragile X syndrome are not simply associated with the presence of cognitive limitations. Compared to males with Down syndrome, the speech of males with fragile X contains more unintelligible strings of syllables, as well as frequent and inappropriate repetition of words, phrases, or topics. Additionally, they more typically talk to themselves, exhibit echolalic speech, and use fewer appropriate nonverbal gestures. Such language patterns are more closely related to those of individuals diagnosed with autism (discussed below). A persistent question evolves around how fragile X syndrome affects the development of

language, as well as how the lack of appropriate language development affects the overall lifestyle of individuals with the syndrome.

Autism and Fragile X Syndrome

Issues in Dual Diagnoses

Researchers have noted the frequent occurrence of hyperactivity, attention deficits, stereotypy, and, at times, self-injurious behaviors among individuals with fragile X syndrome (e.g., Bregman et al., 1987). These characteristics, in addition to the presence of gaze avoidance and communication deficits have provided impetus for investigations of whether a relationship between fragile X syndrome and autism exists. Autism occurs in .04% of the population and is four times more likely to affect males. While the cause of autism remains unknown, some researchers have suggested that autism is due to brain dysfunction (Holmes, 1994). Thus the relationship between these two disabilities remains one worthy of continued study.

Research on the degree of overlap between fragile X and autism has been inconsistent. In 1982, Brown and colleagues reported that 5 of the 27 (18.5%) of their subjects with fragile X had been previously diagnosed with autism. Levitas, Hagerman, Braden, Rimland, McBogg, and Matus (1983) reported that more than 60% of their subjects with fragile X met DSM-III criteria for infantile autism. Similarly, Hagerman, Jackson, Levitas, Rimland, and Braden (1986) reported that 23 of 50 (46%) males in their study had been diagnosed with infantile autism. Conversely, Fryns, Jacobs, Kleczkowska, and Van Den Berghe (1984) reported that, of 21 subjects with fragile X, only 3 (14%) received a diagnosis of autism.

Researchers have also studied individuals who have an initial diagnosis of autism to determine if there is a high prevalence of fragile X among this population. As Bregman et al. (1987) noted, a study in Sweden found that, of 101 subjects with autism, 16% were also diagnosed with fragile X syndrome. Similarly, Brown et al. (1986) screened 183 males identified as autistic and found 24 (13.1%) to be fragile X positive.

Communication Patterns

Due to the general similarity in language development between individuals with fragile X syndrome and those with autism, research has focused on whether qualitative or quantitative language differences exist. Sudhalter, Cohen, Silverman, and Wolf-Schein (1990) examined 33 males in an attempt to investigate possible deviant language patterns. Three conversational contexts were analyzed: direct response, initiation of new material, and topic maintenance. Males with fragile X presented a pattern of deviant language that was qualitatively different from the language exhibited by individuals with Down syndrome, and both qualitatively and quantitatively distinct from language patterns of persons with isolated autism. The latter group generated more deviant language and echolalia when answering questions and initiating communication when compared with those with fragile X, whereas the males with fragile X produced more perseverative deviant language than those with autism. Sudhalter et al (1990) concluded that individuals with fragile X have a distinct pattern of deviant language which differs from the language patterns associated with both autism and other forms of mental retardation. A possible explanation for this deviant repetitive language is symptomatic word retrieval difficulty. The high percentage

of repetitive language could represent a delay tactic used by the subjects while they process others' utterances and search for correct words to use in response.

A related possible source of language difficulties could be perceptual. A number of investigators have noted that males with fragile X who are intellectually disabled have difficulty with auditory reception, auditory sequential memory, and concentration (e.g., Hagerman, Kemper, & Hudson, 1985; Wisniewski et al., 1985). Thus, these difficulties may have linguistic consequences.

Another possible explanation for the deviant language behaviors of males with fragile X syndrome comes from research which indicates that they are more sensitive to social gaze than are males with Down syndrome or isolated autism. Such sensitivity may manifest as repetitive language. Thus, this language pattern may be an indication of social anxiety (Cohen et al., 1988; Sudhalter et al., 1990).

Wolff, Gardner, Paccia, and Lappen (1989) observed the greeting behavior of males with fragile X. It was noted that while greeting both strangers and those with whom they were familiar, 78% of the males in their study turned their head and upper trunk away from the line of gaze, while at the same time accepting and shaking the offered hand. Two of the individuals, both having an additional diagnosis of infantile autism, exhibited greeting behavior that was different from that of the subjects having isolated fragile X syndrome. Indeed, they not only apparently ignored their partner, but also exhibited unrecognizable greeting patterns. None of the subjects with nonspecific mental retardation, and only one of the males with Down syndrome, exhibited such deviant greeting behaviors. Interestingly, males with fragile X under eight years did not manifest the atypical greeting

behavior, whereas most over the age of 12 routinely exhibited the deviant greeting behaviors.

Hypersensitivity

Individuals with fragile X often appear overwhelmed by environmental sounds, sights, movements, and even smells and tastes (King, 1995). These characteristics are similar to those demonstrated by children with autism; researchers have described the problem as an inability to organize sensory input. This hypersensitivity to stimulation may manifest itself in constant talking, hand flapping or biting, avoidance of eye contact, or withdrawal. Hypersensitivity to light touch is usually accompanied by a similar sensitivity to auditory stimuli. When confronted with these stimuli, individuals may try to escape the situation, attempt to counteract the noise by covering their ears, or make loud noises to drown out the incoming stimuli. They may also try to calm themselves by chewing on objects that are readily available, such as shirt or hand, by sucking their thumb, or by rocking. Self-stimulatory behaviors may be a reaction to boredom or to not understanding what is taking place in the environment.

Perspective

Although similarities in behavior exist between fragile X syndrome and autism, there is no conclusive evidence that a relationship between the two conditions exists. While males with fragile X often exhibit autistic-type behaviors, they are usually less significant than the behaviors seen in persons who are clinically diagnosed as autistic. However, the similarity in behavior patterns does lead to difficulty in making a diagnosis of typical autistic-like behaviors versus clinical autism. Past research is often limited in elucidating

the possible relationship exists between autism and fragile X syndrome because: (a) research samples frequently included specific data which cannot be generalized ; (b) varying IQ acts as a confounding factor (i.e., as IQ increases, the number and degree of autistic-type behaviors decrease) (Hagerman et al., 1983); (c) some diagnostic tools may be useful only with individuals in particular settings (Dykens, Leckman, Paul, & Watson, 1988); and, (d) different screening and assessment methods yield varying statistical outcomes (Cohen et al., 1988). However, the significant problem of the past of making an accurate diagnosis of fragile X syndrome was remedied when direct testing for the fragile X gene became a reality. Such testing makes it feasible to test individuals for the presence of the fragile site when they exhibit behaviors associated with autism.

Interventions

Advances in professional knowledge have led to increased possibilities for appropriate interventions for individuals who have been diagnosed with fragile X syndrome. Intervention strategies are typically characterized by a multidisciplinary approach. Plans designed by a team representing medical, educational, and other health professionals (e.g., physical, occupational, speech language therapists) are more likely to achieve success in meeting the special needs of individuals with fragile X, as well as their families.

Medical Interventions

Some of the more common behavior problems associated with fragile X syndrome respond to treatment by medication. Key behavioral problem areas associated with medical interventions are discussed below.

Hagerman (1995) proposed that almost all young boys with fragile X syndrome, and approximately one third of affected girls, exhibit significant attentional problems. Stimulants such as Ritalin and Dexedrine are among the prescribed medications. They are effective in approximately 60 to 70% of the cases of school-aged children through their ability to stimulate the neurotransmitter systems. Thus, these stimulants can result in improvements in attention, hyperactivity, and inhibition.

Clonidine is a common alternative to stimulants for the treatment of hyperactivity. While Hagerman (1995) noted that Clonidine is better than stimulants for calming hyperactive behavior, it does not work as well to improve attention and concentration. It is most effective in calming behaviors when taken in the late afternoon and evening. Sedation is a common side effect.

Moodiness and aggressive outbursts also can be common among adolescents and young adult males with fragile X. These outbursts may be verbal or physical, the latter manifesting as hitting, biting, and kicking behaviors. Mood stabilizers, such as anticonvulsants, can be helpful in improving behavior and decreasing outbursts in individuals who do not have seizures or abnormal EEG patterns. Fluoxetine (Prozac) has been shown to be helpful in decreasing aggression and outburst behavior in approximately 70% of males with fragile X (Hagerman, 1995).

Anxiety and obsessive/compulsive behaviors represent other challenges that have been observed in individuals with fragile X. Benzodiazepines, such as Valium, are effective for anxiety, but they have addictive and sedating qualities. A newer antianxiety medication, buspirone (Buspar), while neither addictive nor sedating, is less effective in

its treatment of panic attacks. Imipramine and Clonidine may also decrease anxiety, although their primary use is in the treatment of hyperactivity. Perseveration in speech and behavior may be helped by serotonin medications.

Although medication may be beneficial in the treatment of many of the behaviors associated with fragile X, it is essential that possible side effects also be considered. Further, the positive response of one individual to a particular medication should not lead professionals to assume that all persons will react in the same manner.

Educational Intervention

According to Wilson, Stackhouse, O'Connor, Scharfenaker, and Hagerman (1994), children with fragile X often demonstrate above-average abilities in the areas of visual memory, sense of humor, hands-on learning, sociability and imitation. The latter two characteristics augment their ability to experience incidental learning, particularly as interaction with non-disabled peers occurs through inclusion.

While specific strengths should be considered in designing educational programs, instruction must also focus on the key challenges faced by students with fragile X. These include organizing and acting on information in a meaningful way, attention focus and span, level of activity, impulsivity, speech and language development, and reducing inappropriate behaviors.

Because students with fragile X often have difficulty with behavioral controls, it is imperative that the context of behavioral problems be included in their education plans. Wilson et al. (1994) recommended O'Neill, Horner, Albin, Storey, and Sprague's (1990) functional assessment guide as a sound tool to determine the function of specific

behaviors. Several guidelines provide direction for identifying the function of undesirable behavior. First, the undesirable behavior must be specifically defined. Second, the environment in which the behavior presents must be identified. Next, behavioral antecedents and functions must be distinguished. Finally, the consequences of the behavior must be identified. Once these steps have been taken, the team of professionals can look for an appropriate alternative behavior that will enable the individual to communicate his/her wants and needs in a positive manner.

Spridigliozzi (1995) outlined some principles for designing an academic curriculum for individuals with fragile X syndrome. Components which Spridigliozzi indicated the curriculum should include are: a reading program that is visually, as opposed to phonetically, based; math lessons which incorporate practical applications; speech/language therapy which emphasizes building language skills and pragmatic communication; and music therapy as a means of teaching academic concepts. Within the classroom, teachers should use visual cues, foster children's imitation skills, and minimize auditory and visual distractions. Spridigliozzi (1995) further recommended developing independent work skills, increasing time on-task through the use of a timer, helping students deal with transition by utilizing a daily schedule complete with pictures, increasing tolerance to change by using pictures as a means of portraying new activities and routines, and implementing a reward system based on daily reports.

Classroom Modifications: Wilson et al. (1994) recommended a number of modifications which allow for a successful classroom experience for students with fragile X syndrome. Optimal seating arrangements include rocking chairs, bean bag chairs, and

chairs which place slight pressure on the base of the spine, while also allowing both feet to fully touch the floor. Seats should be assigned with consideration given to the amount of potential distraction from windows, doors, and in-class noise. A designated quiet area should be available for those times when students need to calm down or relax. Other considerations include minimizing exposure to odors such as perfume and food, the latter due to fragile X children's hypersensitivity to smell.

Children with fragile X often display undesirable behaviors when confronted with changing daily schedules and periods of transition. Providing structure, visual and auditory cues, and pairing the child with fragile X with other students helps alleviate stress and confusion, which may in turn reduce undesirable behaviors. Because they tend to learn routines quickly, providing predictable schedules can also reduce anxiety.

Pictorial schedules are particularly useful in helping students feel in control of their routines and environments. However, one disadvantage of their ability to learn routines quickly is their potential to rely too heavily on the schedule, and thus exhibit undesirable behaviors when the routine in broken. It is suggested that the child be allowed to feel in control of the routine, while components of the routine are gradually changed (Wilson et al., 1994).

Self-Management Skills: As is valid for many students with disabilities, the development of self-esteem, self-efficacy, and a sense of empowerment occurs only as one learns to control his behaviors without relying upon external forces. Thus, self-management skills (e.g., self-monitoring, self-reinforcement), are crucial if goals are to be reached. Self-management provides the means for both structure and maintenance of

control over one's behaviors. Although it may be necessary for professionals and parents to help set goals, the child must share in the process so that ownership and responsibility are achieved. The child must also learn to recognize when target behaviors have occurred, as well as provide self-reinforcement for those behaviors. Finally, the child needs to participate in efforts to adjust goals and review progress made toward achievement of set goals (Wilson et al., 1994).

Curricular Modifications: Curricular modifications and adaptations should take into account the individual interests, abilities, and needs of children with fragile X syndrome (Wilson et al., 1994). Such modifications typically occur in the areas of achievement expectation, activities, and classroom materials. Achievement expectations may be adapted such that the student completes the same activity or uses the same materials as other students but is allowed to do less problems or simpler problems. Activity modifications may include reducing the level of abstraction or difficulty, or developing activities that closely parallel those in the general curriculum. Modification of classroom materials may include highlighting instructions, enlarging text, and using manipulatives. Finally, educators must consider the necessity of age-appropriate, high-interest topics and materials in a curriculum which strives to increase motivation and attention.

It is clear that modifications to the curriculum and environment may be necessary for the student with fragile X to be successful in the general education classroom. However, little research is available to suggest how such modifications can be incorporated into inclusive classrooms. One important issue concerning integrated settings is determining how students with fragile X , whose behaviors often include aggression,

delayed speech, and a varied repertoire of disruptive actions, can be fully included in the daily routine of the general education setting. Students who exhibit extremely disruptive behaviors will have extensive support needs that must be addressed for them to be successful in the general education environment.

Sensory Integration Therapy

Schopmeyer and Lowe (1992) stated that children with fragile X syndrome demonstrate behavioral reactions that are a result of either an overload or a miscommunication of sensory input. Sensory integration deficits can occur in the tactile, vestibular (gravity and movement), proprioceptor (receptors responding to pressure, position, or stretch), visual, auditory, olfactory, and gustatory (taste) systems. These deficits have given rise to consideration of the use of sensory integration therapy, an intervention approach based on the theory that learning is dependent on the ability of individuals to take in sensory information from the environment and to process and integrate this information to plan and organize behavior. If the child has deficits in processing and integrating sensory input, it is hypothesized that conceptual and motor learning can be enhanced by providing expanded sensory intake. For example, a vestibular input calming techniques is placing a child in a suspended swing so that he or she may experience a rhythmic swinging motion.

The assumptions underlying the use of sensory integration therapy have been subjected to a number of research analyses (e.g., Kaplan, Polatajko, Wilson, & Faris, 1993). The controversies surrounding sensory integration therapy stem from the lack of empirical support for its ability to improve students' behavior and academic achievement.

Arendt, MacLean, and Baumeister (1988) concluded that, while evidence indicates that the therapy plays a positive role in the area of motor difficulties, there is no significant difference between the effects of sensory integration therapy as compared to other therapeutic techniques. Further, because sensory integration therapy is used in conjunction with other therapies (e.g., occupational, physical, speech), success may be due to the interaction of these therapeutic techniques. Regardless of the program being used to improve behavior, a reliable form of monitoring should be included as a means of measuring what changes are taking place, as well as the apparent causes for change.

Discussion

Fragile X syndrome is a hereditary chromosomal abnormality with a diversity of implications. There is a wide spectrum of characteristics seen in fragile X syndrome, ranging from physical and behavioral symptoms to speech and language delays. It is associated with cognitive limitations which can include learning disabilities and mild to severe retardation.

Through genetic research, the fragile X gene has been located. Research continues in hopes of finding a way to prevent future families from being affected. Recent media reports of in vivo attempts at the use of fetal gene therapy conducted on animals suggest an exciting future filled with the possible elimination of forms of inherited mental retardation as well as controversy over the ethical implications of cost, availabiltiy, safety, and morality held by society concerning various interventions.

Educational professionals need to focus on the strengths as well as the weaknesses of each person on an individual basis. In allowing each person to develop their fullest

potential with as little restriction to their environment as possible, a large percentage of the individuals with fragile X will be able to become productive members of society. This goal will be realized when support for early intervention and for necessary services is in place, as well as when the general population is educated to accept people who have cognitive limitations. To achieve a positive quality of life for individuals with fragile X, professionals, parents, and friends need to encourage and allow the individual to be an active participant in any decisions being made, and thus encourage a sense of self-empowerment and self-efficacy.

References

Arndt, R.E., MacLean, W.E., & Baumeister, A.A., (1988). Critique of sensory integration therapy and its application in mental retardation. *American Journal on Mental Retardation, 92*, 401-411.

Bodurtha, J., Tams, L., & Jackson-Cook, C. (1992). Prenatal genetic counseling: What is fragile X? Virginia Medical Quarterly, 119, 97-98.

Brown, W.T., Jenkins, E.G., Friedman, E., Brooks, J., Wisniewki, K., Ragatha, S., & French, J. (1982). Autism is associated with the fragile X syndrome. *Journal of Autism and Developmental Disorders, 12*, 303-307.

Brown, W.T., Jenkins, E.C., Cohen, I.L., Fisch, G. S., Wolf-Schein, E.G., Gross, A., Waterhouse, L., Fein, D., Mason-Brothers, A., Ritvo, E., Ruttenberg, B.A., Bentley, W., & Castells, S. (1986). Fragile X and autism. *American Journal on Medical Genetics, 23*, 341-352.

Bregman, J.D., Dykens, E., Watson, M., Ort, S.I., & Leckman, J.F. (1987). Fragile X syndrome: Variability of phenotypic expression. *Journal of the American Academy of Child and Adolescent Psychiatry, 26*, 463-471.

Bregman, J.D., Leckman, J.F., & Ort, S.I. (1988). Fragile X syndrome: Genetic predisposition to psychopathology. *Journal of Autism and Developmental Disorders, 18*, 343-354.

Cohen, I.L., Fisch, G., Sudhalter, V., Wolf-Schein, Hanson, D., Hagerman, R., Jenkins, E.C., & Brown, W.T. (1988). Social gaze, social avoidance, and repetitive behavior in fragile X males: A controlled study. *American Journal of Mental Retardation, 92*, 436-446.

de La Cruz, F.F. (1985). Fragile X syndrome. *American Journal of Mental Deficiency, 90*, 119-123.

Dykens, E., Leckman, J., Paul, R., & Watson, M. (1988). Cognitive, behavioral and adaptive functioning in fragile X and non-fragile X retarded men. *Journal of Autism and Developmental Disorders, 18*, 41-52.

Finucane, B., McConkie-Rosell, A., & Cronister, A. (Eds.). (1993). *Fragile X syndrome: A handbook for families and professionals.* Elwyn, PA: Elwyn.

Fryns, J.P., Jacobs, J., Kleczowska, A., & Van Den Berghe, H. (1984). The psychological profile of the fragile X syndrome. *Clinical Genetics, 25*, 131-134.

Hagerman, R.J. (1995). *Medication intervention in fragile X syndrome* [CD-ROM]. Denver, CO: National Fragile X Foundation.

Hagerman, R.J., Jackson, A.W., Levitas, A., Rimland, B., & Braden, M. (1986). An analysis of autism in fifty males with the fragile X syndrome. *American Journal of Medical Genetics, 23*, 359-374.

Hagerman, R.J., Kemper, M., & Hudson, M. (1985). Learning disabilities and attentional problems in boys with the fragile X syndrome. *American Journal of Diseases of Children, 139*, 674-678.

Hagerman, R.J., Smith, A.C.M., & Mariner, R. (1983). Clinical features of the fragile X syndrome. In R.J. Hagerman & R.M. Boggs (Eds.), *The fragile X syndrome: Diagnosis, biochemistry, and intervention* (pp. 17-53). Dillon, CO: Spectra.

Harvey, J., Judge, C., & Weiner, S (1977). Familial X-linked mental retardation with an X chromosome abnormality. *Journal of Medical Genetics, 14*, 46-50.

Holmes, D.S. (1994). *Abnormal psychology* (2nd. ed.). New York: Harper Collins.

King, L. (1995). *Methods for reducing hypersensitivity to sensory stimulation in autistic individuals.* [CD-ROM] Denver: National Fragile X Foundation.

Levitas, A. (1994). *Behavioral characteristics of the fragile X syndrome.* [CD-ROM] Denver: National Fragile X Foundation.

Levitas, A., Hagerman, R.J., Braden, M., Rimland, B., McBogg, P., & Matus, I. (1983). Autism and the fragile X syndrome. *Developmental and Behavioral Pediatrics, 4*, 151-158.

Lubs, H.A. (1969). A marker X chromosome. American Journal of Human Genetics, 21, 231-244.

Mattei, J.F., Mattei, M.G., Aumeras, C., Auger, M., & Giraud, F. (1981). X-linked mental retardation with the fragile X: A study of 15 families. *Human Genetics, 59*, 281-289.

Newell, K., Sanborn, B., Hagerman, R.J., (1983). Speech and language dysfunction in the fragile X syndrome. In R.J. Hagerman & P. McBogg (Eds.), *The fragile X syndrome* (pp. 175-220). Dillon, CO: Spectra.

Rogers, R. C., & Simensen, R. J. (1987). Fragile X syndrome: A common etiology of mental retardation. American Journal of Mental Deficiency, 91, 445-449.

Rousseau, F., Heitz, D., Biancalana, V., Blumenfield, S., Kretz, C., Boue, J., Tommerup, N., Van Der Hagen, C., DeLozier-Blanchet, C., Croquette, M.F., Gilgendrantz, S., Jalbert, P., Voelckel, M.A., Oberle, I., & Mandel, J.L. (1991). Direct diagnosis by DNA analysis of the fragile X syndrome of mental retardation. *New England Journal of Medicine, 325*, 1673-1681.

Schopmeyer, B. B., & Lowe, F. (Eds.) (1992). *The fragile X child.* San Diego, CA.: Singular.

Spridigliozzi, G.A. (1995). *Potential needs of school-aged children with fragile X syndrome* [CD-ROM]. Denver: National Fragile X Foundation.

Sudhalter V., Cohen, I.L., Silverman, W., & Wolf-Schein, E.,G. (1990). Conversational analyses of males with fragile X, Down syndrome, and autism: Comparison of the emergence of deviant language. *American Journal of Mental Retardation, 94*, 431-441.

Sutherland, G.R., Gedeon, A., Kornman, L., Donnelly, A., Byard, R.W., Mulley, J.C., Kremer, E., Lynch, M., Pritchard, M., Yu, S., & Richards, R. I. (1991). Prenatal diagnosis of fragile X syndrome by direct detection of the unstable DNA sequence. New England Journal of Medicine, 325, 1720-1722.

Turner, G., Brookwell, R., Daniew, A., Selikowitz, M., & Zilibowitz, M. (1980). Heterozygous expression of X-linked mental retardation and the marker X: fra(X) (q27). *New England Journal of Medicine, 303*, 662-664.

Verkerk, A.J.M., Pieretti, M., Sutcliffe, J.S., Fu, Y.H., Kuhl, D.P.A., Pizzuti, A., Reiner, O., Richards, S., Victoria, M.F., Zhang, R., Eussen, B.E., van Ommen, G.J.B., Bolden, L.A.J., Riggins, G.J., Chastain, J.L., Kunst, C.B., Galjaard, H., Caskey, C.T., Nelson, D.L., Oostra, B.A., & Warren, S.T. (1991). Identification of a gene (FMR-1)containing a CGG repeat coincident with a breakpint cluster region exhibiting length variation in fragile X syndrome. *Cell, 65*, 905-914.

Wilson, P., Stackhouse, T., O'Connor, R., Scharfenaker, S., & Hagerman, R. (Eds). (1994). *Issues and strategies for educating children with fragile X syndrome.* Dillon, CO: Spectra.

Wisniewski, K.C., French, J.H., Fernando, S., Brown, W.T., Jenkins, E.C., Friedman, E., Hill, A.L., & Miezejeski, C.M. (1985). The fragile X syndrome: Associated neurological abnormalities and developmental disabilities. *Annals of Neurology, 18*, 665-660.

Wolff, P.H., Gardner, J., Paccia, J., & Lappen, J. (1989). The greeting behavior of fragile X males. *American Journal on Mental Retardation, 93*, 406-411.

Wolf-Schein, E.G., Sudhalter, V., Cohen, I.L., Fisch, G.S., Hanson, D., Pfadt, A.G., Hagerman, R.J., Jenkins, E.C., & Brown, W.T. (1987). Speech-language and the fragile X syndrome: Initial findings. *Journal of the American Speech and Hearing Association, 29*, 35-38.